Case Report

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EXTRAMEDULLARY PLASMACYTOMA OF GINGIVA: A CASE REPORT

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ABSTRACT

Plasmacytoma refers to a malignant monoclonal plasma cell tumor growing within soft tissue. It is similar to multiple myeloma, but lacks the bone changes and resultant hypercalcemia seen in multiple myeloma. Extramedullary plasmacytoma is defined as neoplastic proliferation of plasma cells in soft tissue. It accounts for up to 3% of all plasma cell tumors. Approximately, 90% of extramedullary plasmacytomas are found in the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity. We report a case of extramedullary plasmacytoma of the gingiva.

KEYWORDS: Extramedullary; Gingiva; Plasma cells

INTRODUCTION

Extramedullary plasmacytoma (EMP) is a plasma cell tumor which involves soft tissues, without any signs of systemic spread. It may originate in many sites, although most frequently it occurs in the upper respiratory tract and oral cavity. EMP is present in less than 5% of all plasma cell neoplasms, a percentage similar to that reported for solitary plasmacytoma of bone (SPB).[1-3] Both these tumors are composed of sheets of plasma cells at different stages of maturity, and both respond favorably to appropriate therapy.[4] However, SPB evolves into multiple myeloma (MM) more frequently than EMP.[1,2,5] The etiology of this disease is unknown, but chronic stimulation, overdose irradiation, viruses and gene interactions in the reticuloendothelial system have been suggested as etiologic factors.[2]

CASE REPORT

A 31 year old female patient reported with a history of mass in her lower front gingival region since 3 months (Fig. 1). The mass was asymptomatic and the patient's only concern was cosmetic. Medical history did not reveal any significant concern. A careful drug history revealed that she was on oral contraceptives since 3 months as per her gynecologist's advice. She was advised to undergo In Vitro Fertilisation (IVF) to conceive. She was asked to rule out any dental concerns before undergoing IVF. On intraoral examination the lesion was present as a mass on the gingiva extending from the mandibular right canine to mandibular left canine. It was light purple in colour measuring 4x1.5x1cms in its greatest dimension. The lesion was sessile and surface appeared smooth. The lesion was firm on palpation with absence of bleeding on probing. Radiographic examination did not reveal any bony changes. A complete hemogram was done which showed all blood counts to be within normal limits. Urine examination was normal. An excisional biopsy was done under local anesthesia (Fig. 2). Histopathologic features showed oral mucosa lined by nonkeratinizing stratified squamous epithelium. Areas of ulceration were seen. The underlying connective tissue was infiltrated by closely packed plasma cells arranged in sheets and islands with varying degrees of differentiation, some with perinuclear halo, occasionally 2 nuclei within a single cell. Russell bodies were seen. Our diagnosis was plasmacytoma (Fig. 3). The patient was followed up for one year without any recurrence.

DISCUSSION

Plasma cells are medium sized round to oval cells with eccentrically placed nuclei. The cells range in size from 10 to 20µm and the nuclear cytoplasmic ratio is 1:2. The nuclear chromatin is often arranged in a cartwheel-like or clock-face pattern. Plasma cells are usually found in the red pulp of spleen, tonsils, medulla of the lymph nodes, nasal mucosa, upper airway, lamina

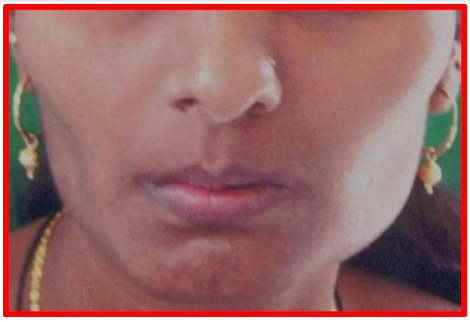

Fig. 1: Facial Photograph showing absence of extraoral swelling

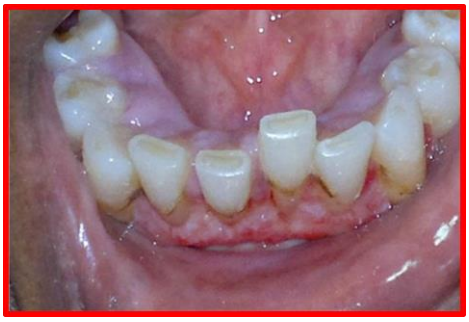

Fig. 2: Postoperative Intraoral Photograph after excision of the lesion

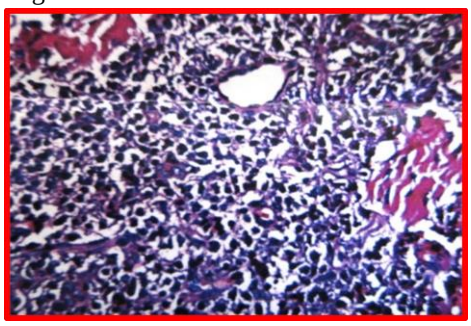

Fig. 3: Closely packed plasma cells with perinuclear halo

propria of the gastrointestinal tracts and sites of inflammation. Their main function is to produce immunoglobulins or antibodies.[6,7] Solitary plasmacytoma of bone (SPB) and extramedullary plasmacytoma (EMP) are unusual solitary tumors of plasma cell origin. They represent distinct manifestations of a disease continuum, whereby the clinical findings are critical to diagnosis. Plasma cell neoplasms are histologically similar, and distinguishing one from the other has significant implications for treatment and survival. Plasma cell neoplasms are relatively unusual malignancies of the head and neck region. Clinical manifestations of plasmacytoma of the oral cavity consist of jaw pain, tooth pain, paresthesia, swelling, tissue mass, mobility of teeth, migration of teeth, hemorrhage and pathologic fracture of the involved bone.[8-10] According to Miller, EMP of the oral cavity can show a cauliflower growth. Solitary plasmacytoma differs from MM by lack of plasma cell infiltration in a random bone marrow biopsy. In patients with EPM, the blood counts are within normal limits, with no signs of anaemia, hypercalcemia and renal failure. Histopathologic features of EMP show a connective tissue greatly infiltrated by plasma cells which are like focal sheets, small islands, or plasmacytoid nodules.[8] The conversion rate of EMP to MM is 15 –20%. The conversion rate of EMP to SPB is 48% and is associated with a poorer prognosis.[10] Harwood *et al*., reported the increased rate of conversion to MM if the EMP involved the adjacent bone. Local recurrence has been reported to be up to 10%.[10] Dissemination of the tumor takes place in 35 – 50% of EMPs. The differential diagnoses of EMP are plasma cell granuloma, pseudolymphoma, and reactive plasmacytic hyperplasia.

CONCLUSION

The cure of the disease can be achieved by radiotherapy in almost all cases. Surgical removal of the tumor can be performed, at diagnosis, as sole treatment of small masses and also when local irradiation has not been successful in eliminating the mass, as in the case of a local deposit of amyloid substance or residual disease. Treatment with chemotherapy does not appear to be indicated because it had no effect on the course of EMP, while disease dissemination can be successfully treated with alklyating agents.

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